Intradural Extramedullary Spinal Tumors

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Pathology

* most are slow-growing and benign (vs. epidural tumors).
* usually involve only few spinal segments.
* if tumor occludes spinal arteries → myelomalacia.

Frequency

1. **Neurofibromas**
2. **Schwannomas** (Schwann cells form covering of spinal nerve roots)
3. **Meningiomas**
4. Other rare cases - **lipoma**, **dermoid**, **epidermoid**, **teratoma**.
5. **Metastases** (relatively uncommon):
   1. leptomeningeal metastases of systemic tumors (esp. breast carcinoma, melanoma, leukemia, lymphoma)
   2. drop metastases of intracranial tumors (e.g. ependymoma, medulloblastoma).

Clinical Features

myelopathy & radiculopathy - Extramedullary Cord / Root Compression [see p. Spin1 >>](http://www.neurosurgeryresident.net/Spin.%20Spinal%20Disorders\Spin1.%20GENERAL%20-%20Spinal%20Syndromes.pdf#Extramedullary_Cord_Compression)

* initial symptoms of ***nerve sheath tumors*** – prominent focal pain and paresthesias (involvement of dorsal roots). *i.e. tend to cause radiculopathy before becoming large enough to cause myelopathy*
* initial symptoms of ***meningiomas*** – myelopathy (radiculopathy develops later).
* compression first interrupts functions of pathways that lie at periphery of spinal cord.
* eccentrically placed tumors may cause typical *Brown-Sequard syndrome*.
* if untreated → complete loss of function below level of lesion (complete spinal cord transection).
* large cervical tumors (esp. with extradural component) may be palpated.

Diagnosis

**Plain X-ray** - occasionally enlarged intervertebral foramen, localized widening of spinal canal.

* *calcification* is rare (heavily calcified intraspinal mass is usually extruded disc material).

**MRI with gadolinium** - enhance brightly.

* features of ***nerve sheath tumors*** - prominent CT/MRI enhancement, characteristic tumor extension through enlarged neural foramen!

**Myelo****graphy**:

1. *filling defect in subarachnoid space* (meniscus corresponding to outline of tumor)
2. *filling defect displaces cord away from ipsilateral dura* → widening ipsilateral subarachnoid space while narrowing contralateral space; plane of cleavage is usually visible between tumor and spinal cord.

(vs. intramedullary tumors - fusiform cord enlargement and circumferential narrowing of adjacent subarachnoid space; extradural tumors - displace cord, but result in narrowing of both ipsilateral and contralateral subarachnoid spaces)

* *leptomeningeal metastases* - small nodules attached to surface of cord or cauda equina nerve roots.

**CSF** – cytology is key diagnostic test for *leptomeningeal metastases*; for other tumors, LP is not diagnostic (and relatively contraindicated)!

* in complete subarachnoid block, CSF is xanthochromic (due to high protein content); some tumors secrete large amounts of proteins (values > 1000 mg/dl may lead to communicating hydrocephalus → brain dysfunction!).
* slight pleocytosis in 30% patients.
* glucose content is normal (unless tumor of meninges is present).

**Angiography** is performed for surgical planning if tumor is in lower thoracic region – ***to locate artery of Adamkiewicz*** – must be preserved during surgery (supplies spinal cord!).

Treatment

Spinal cord compression – see [p. Onc56 >>](http://www.neurosurgeryresident.net/Onc.%20Oncology\Onc56.%2520Extradural%2520Spinal%2520Tumors,%2520Vertebral%2520Tumors.pdf#Spinal_cord_compression_treatment)

**Total surgical excision** is main treatment for *benign tumors*.

* **steroids** in perioperative period.
* laminectomy is adequate approach (certain tumors require lateral or even anterior approach).
* *extraspinal tumor extensions* can be removed by extending initial laminectomy exposure laterally (some may require separate operation through thoracotomy, costotransversectomy, or retroperitoneal approach).
* sectioning of dentate ligament ensures access to tumor; dorsal roots can also be sectioned.
* excise involved portion of **dura** (*meningioma*) or involved **nerve root**\* (*neurofibroma*, *schwannoma*). \*frequently nonfunctional at time of surgery
* after extensive surgery, ***spinal deformities*** (which may have been present preoperatively) may appear or increase; H: fixation, laminoplasty.

*Leptomeningeal Metastases* are not treated surgically! *see below* [>>](#LEPTOMENINGEAL_METASTASES)

Specific Tumor Types

Meningioma

[also see p. Onc38 >>](http://www.neurosurgeryresident.net/Onc.%20Oncology\Onc38.%20Meningioma.pdf)

* 70-80% located in *thoracic cord*, 10-20% in cervical spine, 3% in lumbar spine, 2% in foramen magnum.
* 5-10 times more frequent in women than in men.
* 7% meningiomas are purely extradural, 6% - intradural and extradural.

N.B. lesion with both intradural and extradural components is most likely to be nerve sheath tumor!

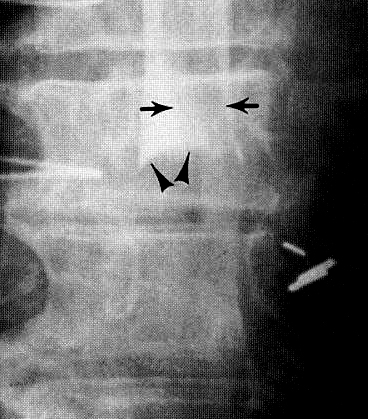
* 2% are multiple (associated with neurofibromatosis-2).
* typically ***globoid***.
* myelopathy > radiculopathy; high incidence of Brown-Sequard syndrome (> 50% meningiomas are positioned laterally).
* **MRI** - broad-based attachment to dura.
* **dural base** should be resected if it is easily accessible (if not → cauterize and scrape with microdissector).
* very high 5-year survival rate - 99.5%.

*A.* T1-MRI - sharply circumscribed low-intensity mass (*arrows*).

*B.* Contrast T1-MRI - intense enhancement of well-circumscribed extramedullary mass (*arrows*) which displaces spinal cord to left, widening cistern adjacent to mass:

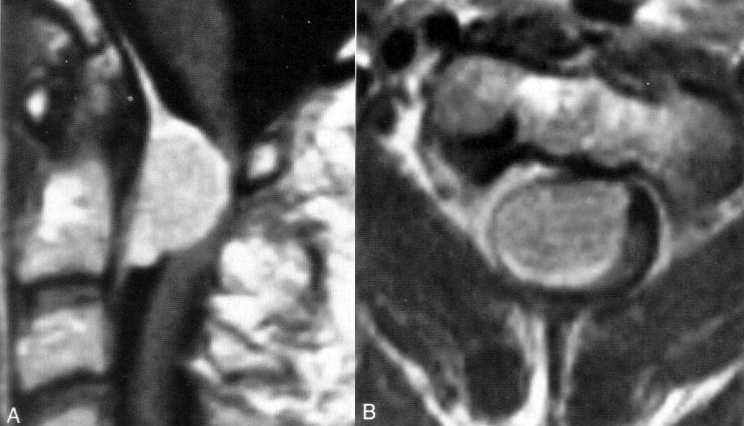


AP thoracic myelogram (via C1-2 puncture) - leftward displacement of cord (*arrows*), widening of ipsilateral subarachnoid space, narrowing of contralateral space, and rounded intradural filling defect (*arrowheads*):

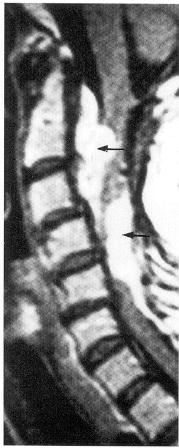


A. Contrast T1-MRI - large intradural mass at C1-2.

B. Contrast T1-MRI - large meningioma, marked spinal cord compression.

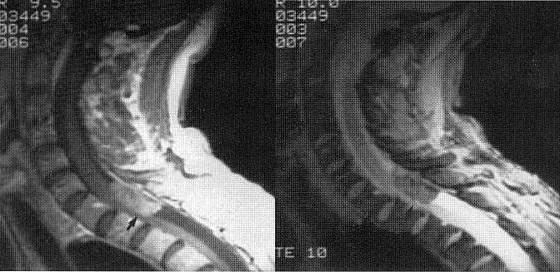


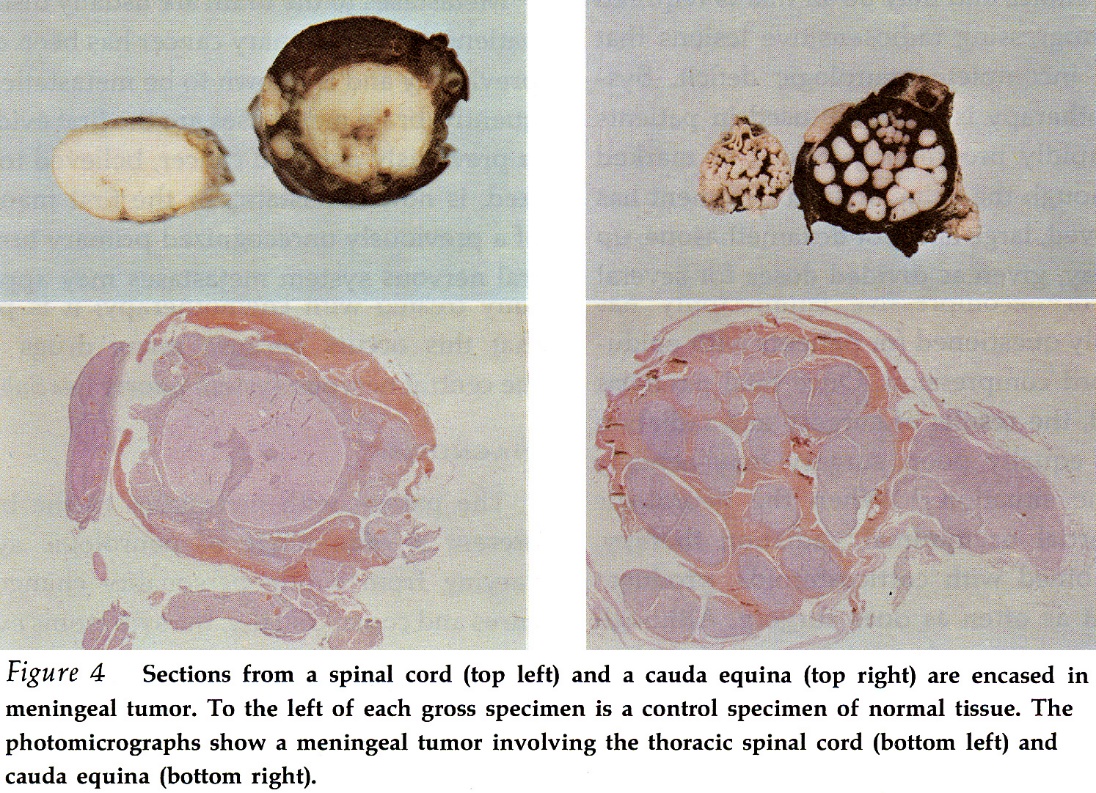
Recurrent cervical meningioma (contrast T1-MRI) - extensive intradural tumor (white signal - *black arrows*) partly surrounding spinal cord:



A) Contrast T1-MRI - uniformly enhancing, dorsal intradural extramedullary mass compressing cord at cervicothoracic junction.

B) T2-MRI - low signal intensity within mass, common in meningiomas:



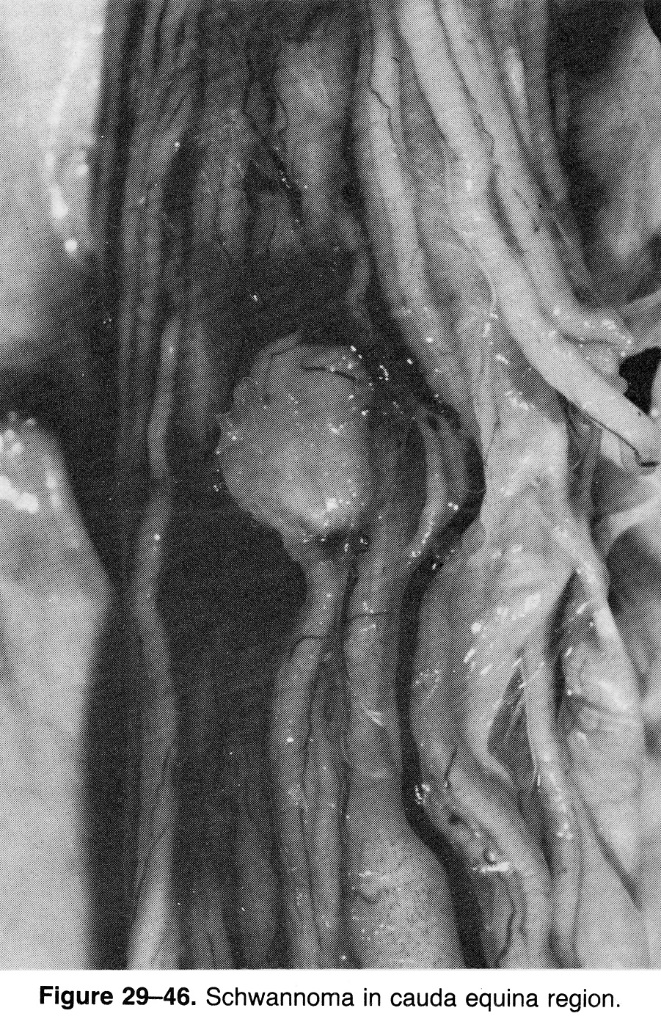


Lipoma

* as lesions of filum terminale, spinal cord, or lipomeningomyeloceles.
* contain variable amounts of fibrous connective tissue (often adherent to cord, meninges, or cauda equina).
* CT - low-density fatty mass with mass effect on adjacent cord.
* bright on T1 and dark on T2.

Neurofibroma, Schwannoma

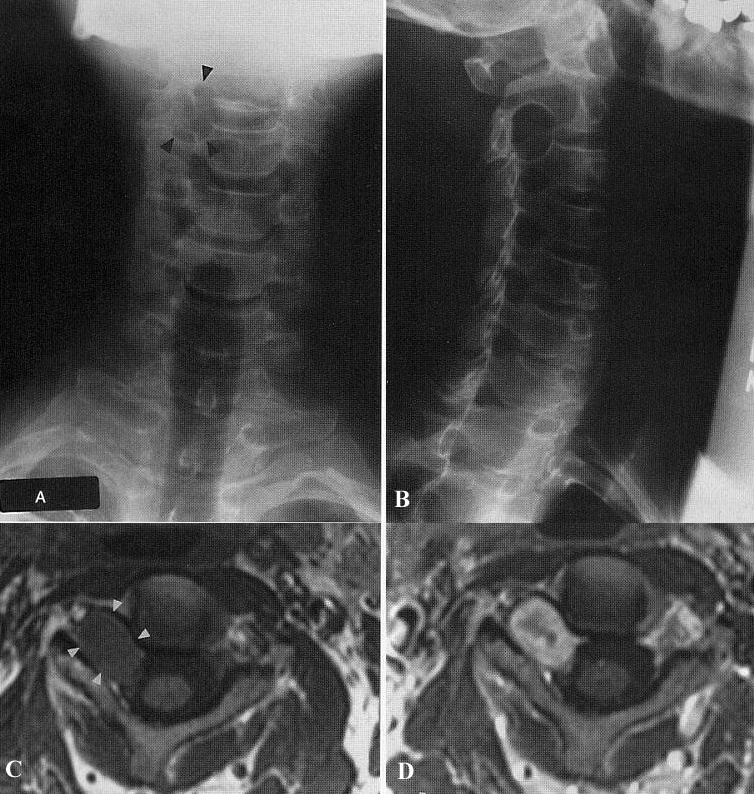
* benign tumors of nerve sheath.
* typically arise from *dorsal roots* - radicular pain is often first symptom (radiculopathy → ≈ 2 years → myelopathy).
* *neurofibromas* - often fusiform and multiple (esp. in neurofibromatosis); *schwannomas* - tend to be solitary and arise eccentrically from nerve sheath.
* 30% have extradural component (“hourglass” or “dumbbell” shape)
* prominent CT/MRI enhancement.
* CSF protein is elevated (because of protein secretion by tumor) vs. *meningiomas*.
* surgical resection.



Right C3 neurofibroma:

A, B (AP X-ray) - apparent enlargement of right C2-C3 nerve root foramen (*arrowheads*); preservation of sharp cortical margins indicates slow growth.

C, D (precontrast T1-MRI, contrast T1-MRI) - largely extradural, densely enhancing soft-tissue mass within right C2-C3 foramen (*arrowheads*); only small component of tumor is intradural:

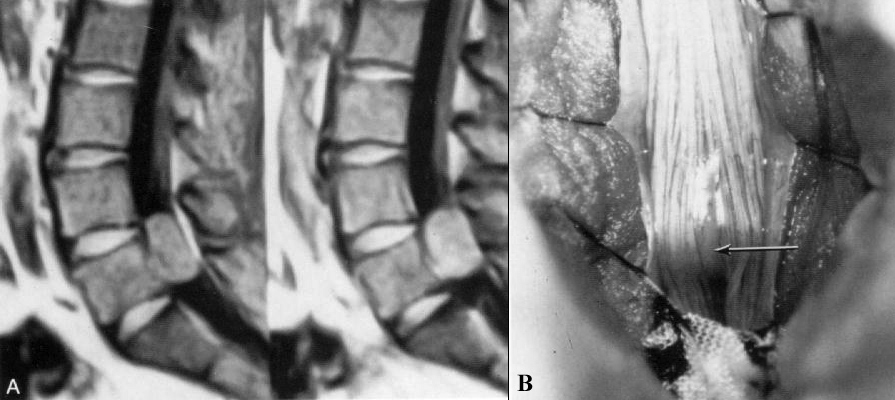


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| Neurofibromatosis (CT myelography) - two neurofibromas, mainly intradural, distend thecal envelope, severely compressing spinal cord (*arrow*):  D:\Viktoro\Neuroscience\Onc. Oncology\00. Pictures\Spinal neurofibromas (CT).jpg | Neurofibroma (CT myelogram) - pronounced posterolateral displacement and compression of spinal cord (crescentic filling defect surrounded by contrast medium - *arrowheads*); large round filling defect:  D:\Viktoro\Neuroscience\Onc. Oncology\00. Pictures\Cervical Neurofibroma (CT myelogram).jpg |

Schwannoma:

A. Contrast T1-MRI - large intradural tumor at L5 level.

B. Intraoperative photograph - large intradural schwannoma (*arrow*) that displaces cauda equina dorsally.



Leptomeningeal metastases

* spinal cord compression develops rapidly.
* frequently multiple.
* CSF - malignant cells, glucose↓, protein↑.
* treatment (complete surgical removal is almost never possible):
  1. **radiotherapy** + **chemotherapy**
  2. **radiotherapy** to entire neuraxis

|  |  |
| --- | --- |
| Subarachnoid "drop metastases" of cranial malignancy (myelography, frontal view): one large, rounded, intradural filling defect (*arrows*) as well as more subtle nodular enlargement of nerve roots in cauda equina:  D:\Viktoro\Neuroscience\Onc. Oncology\00. Pictures\Spinal metastasis (myelography).jpg | Intradural metastases (contrast T1-MRI):  A) Metastatic medulloblastoma - tumor aggregation in termination of caudal sac (*arrows*), and coating of spinal cord.  B) Cord encasement by metastatic breast carcinoma (*arrow*).  D:\Viktoro\Neuroscience\Onc. Oncology\00. Pictures\Intradural metastases (MRI).jpg |

Spinal Cysts

- rare congenital, developmental mass lesions (intramedullary or extramedullary intradural).

* found pre­dominantly in cervical and thoracic regions.
* present clinically by 4-5th decade of life.
  1. *Arachnoid cyst* - most-common intraspinal cyst; single-layered arachnoid cell lining, without epithe­lium or cilia.
  2. *Ependymal cyst* has ciliated, cuboidal, or columnar epithelial lining.
  3. *Enterogenous cyst* (derived from neurenteric canal or primitive endoderm) lined by ciliated, secre­tory columnar epithelium and can produce mucin; may be associated with GI tract dupli­cation and dysraphic vertebral abnormalities. [also see p. Onc30 >>](http://www.neurosurgeryresident.net/Onc.%20Oncology\Onc30.%20Dermoid,%20Epidermoid,%20Cysts,%20Lipoma.pdf#NEURENTERIC_CYST)
  4. *Branchiogenic cyst* has respiratory epithelial lining and congenital vertebral anomalies in thoracic spine as well.
* treatment:
  + 1. micro­surgical excision
    2. cyst fenestration
    3. cysto-subarachnoid, cysto-pleural, or cysto-peritoneal shunting.

Bibliography for ch. “Neuro-Oncology” → follow this [link >>](http://www.neurosurgeryresident.net/Onc.%20Oncology\Onc.%20Bibliography.pdf)

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